
Multiple Enchondromatosis: Ollier's disease- A case report.

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Citation

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Abstract

Multiple enchondromatosis is a rare developmental disorder characterised by large rounded masses or columns of cartilage in the metaphysis of certain bones, particularly the long bones¹. It is defined by the presence of at least three enchondromas. It is characterised by asymmetric distribution of lesions and an extreme clinical variability. Multiple enchondromatosis may cause devastating disfigurement and even adversely affect the growth and function of an extremity. Radiographs show typical changes and are generally diagnostic. There is no medical treatment for enchondromatosis. Surgical intervention is indicated in case of complications (pathological fractures, growth defect, malignant transformation). We hereby report a case of multiple enchondromatosis in an 8 yrs old child with growth disturbances.

CASE REPORT

A 8 year old female patient presented to us with 1 year history of progressively increasing painless swelling in the

left knee. No other medical problems were reported. On physical examination there was broadening of left knee with shortening of left leg by 2cm (Fig.1).

Figure 1

Figure 1: Clinical photograph of a patient showing broadening of left knee with shortening of the same leg.



Figure 2

Figure 2: Anteroposterior and lateral radiograph of knee and ankle showing typical lesions of enchondromatosis.



On further radiological evaluation similar lesions were also found involving the proximal and distal femur with involvement of ilium also on the ipsilateral side although there was no apparent swelling in these areas (Fig.3).

There were no other significant findings. On radiological evaluation there were multiple, radiolucent, homogenous lesions with oval or elongated border and well defined slightly thickened bony margin involving the proximal and distal tibia. These lesions resulted in metaphyseal widening around the knee and were typical of multiple enchondromatosis (Fig.2).

Figure 3

Figure 3: Radiograph of pelvis and thigh with typical lesions of enchondromatosis in proximal and distal parts of femur and changes in illium on ipsilateral side.



A family history of similar disorder could not elicited.

The patient was counselled about the nature of the disease, its complications and was given various surgical options and the importance of continuous follow up. Since patient was not having much problem clinically he did not agreed for any surgical options and was discharged on request. Patient was given full advice and importance of follow up at the time of discharge. Presently patient is under our continuous followup.

DISCUSSION

Enchondromas are common benign usually asymptomatic cartilage tumours which develop in the metaphysis and may become incorporated into the diaphysis of long tubular bones, in close proximity to the growth plate cartilage²³⁴. Typically the tumour involves the short bones of hands and feet⁵. Tumours may be limited to single bone (solitary) or

may involve multiple bones (multiple enchondromatosis). Multiple enchondromatosis with a predominantly unilateral distribution is classically referred as Ollier's disease. The association of multiple enchondromas with soft tissue haemangioma is known as Maffucci's syndrome⁶. If tubular bones of the hands are involved, painless swelling of fingers may be the initial complaint⁷. The swelling may be severe enough to impair function.

Malignant sarcomatous transformation is a common feature in case of multiple enchondromas and has been reported in approximately 50% of cases⁵⁸. Malignant transformation is clinically signalled by increased size of the tumour and pain during adult life. Biopsy is needed to demonstrate the sarcomatous transformation⁸⁹. Malignancy is higher in maffucci's syndrome, the prognosis of which is more severe than ollier's disease²³ and lower in hereditary multiple exostosis. Association of ollier's disease with other tumours have been reported³¹⁰¹¹¹².

Enchondromas have a typical appearance radiographically. The radiographic findings are typically more severe than the clinical examination examination would suggest. Long bone lesions appear as longitudinal, elongated radiolucent streaks in the metaphysis extending to the shaft. In young children, the epiphysis is not involved. Lesions may extend into epiphysis at later ages after the epiphysis has closed⁷.

Solitary lesions are treated with curettage and bone grafting⁵. The recurrence rate may be high after curettage¹³. In some cases, enchondromas may be replaced by mature bone without any sequelae⁸. The treatment of multiple enchondromas should be tailored to the distribution of lesions involving the most severely deformed extremities. Epiphysiodesis or limb lengthening procedures may be required to resolve limb length discrepancies⁵.

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